

CASE REPORT: PERSISTENT PROATLANTAL INTERSEGMENTAL ARTERY¹*Jehad Al-Habsi, MD and ²Sharad S. Rajamani, MCh, FICS, FAVNS, FEBNS, PhD¹College of Medicine and Health Sciences, Sultan Qaboos University, Muscat, Sultanate of Oman.²Neurosurgery Department, Khoula Hospital, Muscat, Sultanate of Oman.***Corresponding Author: Dr. Jehad Al-Habsi**

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ABSTRACT

Background: Remnants of fetal carotid-basilar circulation are rarely identified. Usually, they are asymptomatic and incidentally found on imaging. A case of proatlantal intersegmental artery diagnosed during angiography for an arteriovenous malformation is described. History, embryological development, and anatomy of the proatlantal intersegmental artery are reviewed. **Case Report:** A 13-year-old boy was brought to the emergency services with a history of sudden onset headache associated with loss of consciousness and then vomiting. Computed tomography (CT) scan showed intraventricular bleeding in the lateral ventricles extending to the fourth ventricle associated with dilatation. Digital Subtraction Angiography (DSA) showed an AVM in the temporal periventricular region, Spetzler Martin grade 3. The patient underwent embolization in the same sittings. Incidentally, It was found a shunting of blood from the ascending pharyngeal to the vertebral artery at the CV Junction was seen. This is representing a type 2 PPIA. **Conclusions:** Most proatlantal intersegmental arteries are found incidentally. Understanding anomalous vascular anatomy such as persistent carotid-vertebrobasilar anastomoses, and the associated anatomical variations, is very essential in planning head and neck surgeries.

KEYWORDS: Persistent proatlantal intersegmental artery, proatlantal intersegmental artery, neurosurgery, anatomy, case report, vascular.

INTRODUCTION

There is a discrepancy in the literature about the historical facts regarding the proatlantal intersegmental artery. As per the review done by Ljiljana et al, this primitive vessel was observed and interpreted in the bovine embryo by Fropier and in the rabbit embryo by Hochstetter. proatlantal intersegmental artery was interpreted by Schmeidel in a human embryo.^[1] On the other hand, it was reported that Gottschau was the first one who described this carotid-vertebral anastomosis in humans.^[1,2]

Persistent proatlantal intersegmental artery (PPIA)

There is a discrepancy in literature when it comes to the first description of PPIA. It was pointed out that Congdon was the first to describe PPIA, while Pasco et al.^[3] assign its description to Gottschau. However, the first angiographical report of PPIA was given in 1966.^[2-4]

Case History

A 13-year-old boy was brought to the emergency services with a history of sudden onset headache associated with loss of consciousness and then vomiting. Computed tomography (CT) scan showed intraventricular bleeding in the lateral ventricles extending to the fourth ventricle associated with

dilatation. There was a casting of blood in the lateral ventricle. CT angiography was suggestive of an arteriovenous malformation (AVM) in the right temporal region, involving the wall of the temporal horn of the right lateral ventricle.

Digital Subtraction Angiography (DSA) showed an AVM in the temporal periventricular region, Spetzler Martin grade 3. The patient underwent embolization in the same sittings.

Incidentally, It was found that extracranial circulation had a shunt from the external carotid artery to the vertebral artery on the left side as demonstrated in [fig 1]. The left vertebral artery was not originating at its normal origin. however, during injection of the left ECA, shunting of blood from the ascending pharyngeal to the vertebral artery at the CV Junction was seen. This is representing a type 2 PPIA. The patient provided informed consent to the publication of this case.

DISCUSSION**Embryology**

During early embryogenesis carotid-basilar anastomoses and primitive carotid vertebral are formed at approximately 24days. Third aortic arches and the ventral aorta supply blood to the primitive internal

carotid artery (ICA) until 24-28 days. The hindbrain circulation develops from two longitudinal neural plexuses merging in the midline to form the basilar artery (BA). It is demonstrated in [Fig 2] that longitudinal neural plexus is supplied by the cranial extension of ICA and below by the cervical intersegmental arteries and four transient anastomotic channels. These channels are trigeminal, optic, hypoglossal, and proatlantal intersegmental arteries which connect the hindbrain vascular plexus and anterior carotid circulation. [Fig 3] is a schematic presentation of carotid-basilar anastomoses.

In 28-30 days of embryo life (5-6mm) transient anastomotic channels provide proximal supply to longitudinal neural arteries via primitive proatlantal intersegmental arteries and cervical carotid-vertebral anastomoses. These anastomotic channels start to involute when the embryo is 7-12mm. The proatlantal intersegmental artery (PIA) is the last one to involute and it completely disappears at about 12-14mm. By the end of the sixth gestational week, PIA normally regresses to form the intracranial vertebral artery.^[1,5-8]

Anatomy

When PPIA presents unilaterally, it originates from ICA at the C2 vertebra level, or C3 vertebra, or at the level of C2-C3, or C4 vertebra. In one case it was reported that PPIA begins 2 cm above the origin of the left ICA.^[11]

Bilaterally, it was found the PPIA originated from ICA at the level of the C2 vertebra. This artery usually has a horizontal course along the posterior arch of the atlas or it has a posterior curve before joining the Atlantic part of VA (V3).^[9] Pasco *et al.*^[3] reported a common trunk originating from the carotid bulb giving two branches: anterior ICA and a posterior branch called pseudo vertebral artery. PPIA is usually found lateral to the ICA; it curves dorsally and above the atlas entering through the foramen magnum into the cranial cavity.^[3]

PPIA can sometimes originate from the External Carotid Artery (ECA) near its origin, or at the level of the atlas when it shares the origin of the occipital artery. It can originate from ECA at the level of C2, C3-C4, and C4.^[11]

Finally, PPIA can originate from the common carotid artery either unilaterally or bilaterally. Palmer and Philips described an abnormal artery that arose from the left common carotid bifurcation and passed upwards obliquely between atlas and occiput similarly to the course of a normal VA. This artery was named PPIA.^[7]

Types

Type 1

It is assumed that type 1 is the true PPIA. It begins from the ECA, the ICA, or the CCA and it joins the intracranial part of VA after passing through the foramen magnum without passing through the transverse foramina of cervical vertebrae [Fig 4a-c].^[7] Kohei

Ishikawa *et al* reported a case of a patient who presented with dizziness. Computed tomography angiography revealed a type 1 PPIA associated with segmental dysplasia of the right internal carotid artery and Aberrant right subclavian artery. It emphasized the importance of the knowledge of anatomical variations. It is very essential for interventions such as angiography or endovascular therapy and the clinical follow-up¹¹. Similarly, Chuan-Min Lin *et al* agree on the importance of the knowledge of anatomical variations including PPIA in clinical management. Chuan-Min Lin *et al* summarized 16 cases of PPIA with VA stenosis; six of the cases had type 2 PPIA, nine cases of type 1 PPIA, and a case of mixed PPIA. Cases were compared in a table including the authors, year of publication, clinical presentation, and management. Management was tailored for every case documented. the management options, which were reported, included stent, Endarterectomy, angioplasty, and Wingspan stent.^[11]

Type 2

It is postulated that type 2 is a persistent primitive first cervical intersegmental artery. Type 2 is arising from the ECA and runs posteriorly which is similar to the course on the occipital artery. It passes through the transverse foramen of C1, then after passing through the foramen magnum it merges with V3 of the VA artery [Fig 4d]. It is located more lateral compared to type 1.^[10,12,13] PPIA might be associated with different variants, and complications and can present with a different presentation. Akira Uchino and Kazuo Tokushige reported a case of a small type 2 left proatlantal artery with normal ipsilateral vertebral artery and aberrant right subclavian artery associated with a bi-carotid trunk which is reported as the first case of its type in the relevant English language literature. It was documented that the previous case was of an elderly patient who presented with cerebral infarction in the right basal ganglia.^[13] On the other hand, an adult patient with vertigo was found to have an absent bilateral vertebral artery on neck ultrasound. MR angiography revealed bilateral type II PIA arose from the bilateral cervical external carotid artery.^[14] Furthermore, A case of type II PIA with vertebral artery hypoplasia with ipsilateral intracranial vertebral artery stenosis and contralateral vertebral artery occlusion. The patient was found to have multiple infarctions over pons, bilateral occipital lobes, and cerebellums were reported.^[11] Gagandeep Choudhary and his team summarized previously reported twenty-one cases of type II PIA in a table where the author, sex of the patient, the side of type II PIA and Additional anomalies were detailed. Some of the associated anomalies mentioned Vein of Galen malformation, Aneurysm at the MCA trifurcation, AVM fed by left PCA, Aneurysm at the MCA trifurcation, and Saccular aneurysm of the left ICA.^[10]



(Fig 1A)



(Fig 1B)

Fig 1: Angiography of left external carotid artery of a 13-year-old boy with sudden onset headache associated with loss of consciousness and then vomiting showing (Fig 1A) anterior posterior view of external carotid artery circulation demonstrating a shunt (arrow) from external carotid artery to vertebral artery on the left side of extracranial circulation and a (Fig 1B) lateral view.

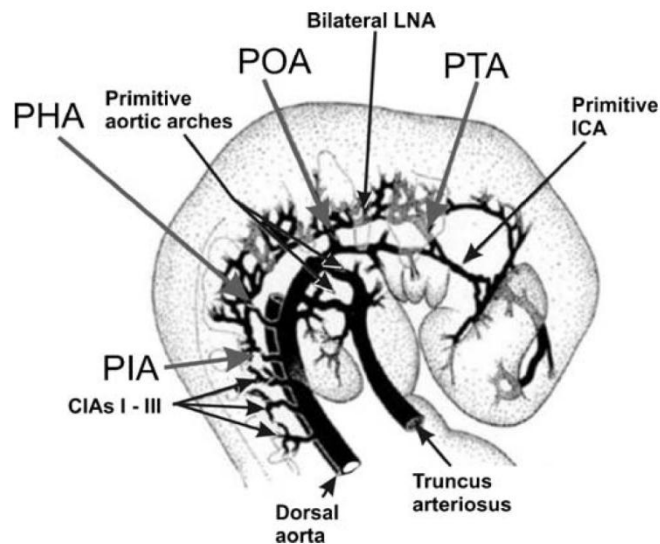


Fig 2: Modified drawing⁷ shows cranial arteries at 5 weeks of gestation demonstrating pre-segmental arteries connecting primitive ICA with bilateral LNA plexus.

ICA= internal carotid artery; CIAs I–III= cervical intersegmental arteries I–III; LNA= longitudinal neural artery; PHA= primitive hypoglossal artery; PIA= primitive proatlantal intersegmental artery; POA= primitive optic artery; PTA= primitive trigeminal artery.

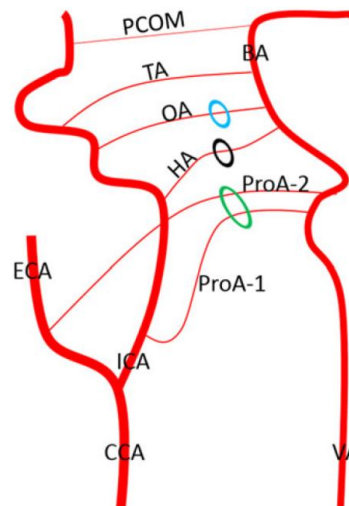


Fig 3: Diagram¹⁰ demonstrating carotid-basilar anastomoses from cranial to caudal: posterior communicating artery (PCOM), trigeminal artery (TA), optic artery (OA), hypoglossal artery (HA), proatlantal artery type 2 (ProA-2), and proatlantal artery type 1 (ProA-1). The blue circle indicates the internal auditory canal; black circle, hypoglossal canal; and green circle, the foramen magnum. BA= basilar artery; ECA= external carotid artery; ICA= internal carotid artery; VA= vertebral artery.

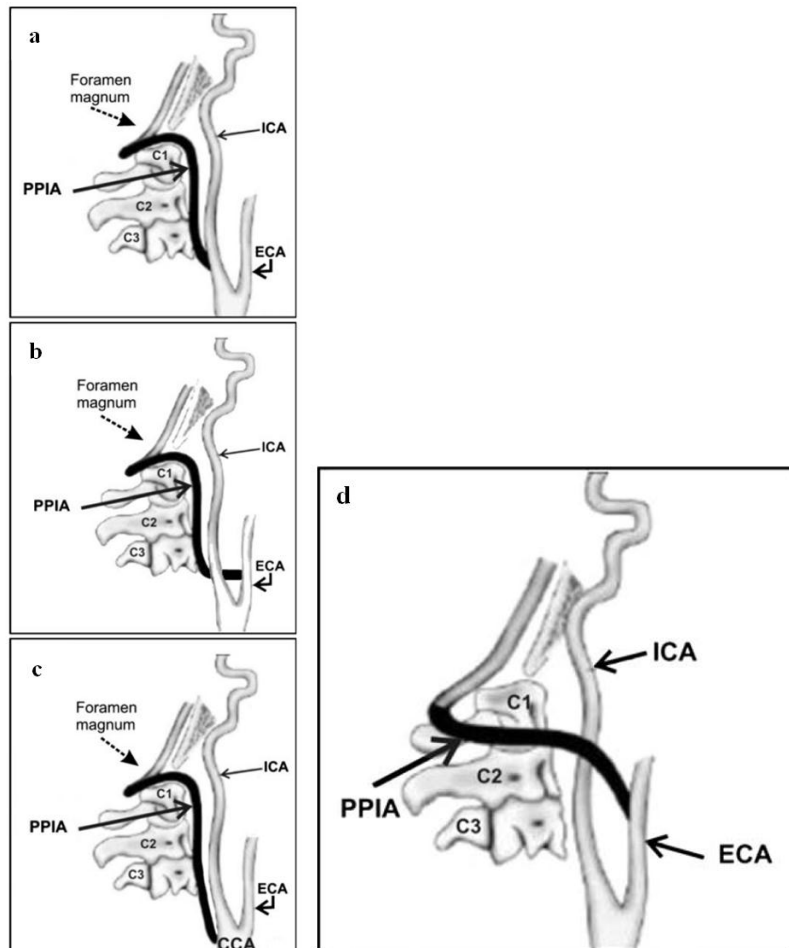


Fig 4: Modified drawing of the first type of PPIA with its origin either from the internal carotid artery (ICA) (a) or external carotid artery (ECA) (b) or from the common carotid artery (CCA) (c). Modified drawing of the second type of Persistent proatlantal intersegmental artery (PPIA) (d) with its origin from the external carotid artery (ECA) and its passage through the foramen transversarium of the C1 vertebra. ICA= internal carotid artery; C1–C3= first three cervical vertebrae

CONCLUSION

Although, most Proatlantal intersegmental arteries are found incidentally. Understanding anomalous vascular anatomy such as persistent carotid-vertebrobasilar anastomoses, and the associated anatomical variations, is very essential in planning head and neck surgeries. Recognition of the PPIA is extremely important in case of surgical ligation or embolization of the external carotid artery is planned.

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